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Benign Schwannoma of the Liver: A Case Report

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Abstract

Neurogenic tumors of the liver are very rare, irrespective of associated neurofibromatosis. We report here a well-documented case of benign schwannoma in a 56-year-old woman without neurofibromatosis, including imaging and pathological examinations.

Introduction

Tumors and tumorlike condition of the peripheral nerves are classified into the following four categories: (i) neuroma, a benign nonneoplastic overgrowth of nerve fibers and Schwann cell; (ii) schwannoma (neurilemmoma) and (iii) neurofibroma, two benign neoplasms; and (iv) malignant schwannoma (malignant peripheral nerve sheath tumor)^{1,2)}. Benign schwannoma is an encapsulated neoplasm, containing cystic areas, especially in a large tumor. Its microscopic appearance is so distinctive as to be easily distinguished from other neurogenic tumors. The common locations of benign schwannoma are the flexor surfaces of the extremities, neck, mediastinum, retroperitoneum, posterior spinal roots and cerebellopontine angle¹⁾. It is rarely found in the hepatobiliary system. We report a benign schwannoma of the liver in a 56-year-old woman who was preoperatively diagnosed as cystadenocarcinoma of the liver.

Case Report

The patient was a 56-year-old woman who initially complained of compression sense in the epigastric and anterior chest area. Physical examination was normal and no cafe-au-lait spots or cutaneous neurofibromas were found. Ultrasonography (US), computed tomography (CT) (Fig. 1), and magnetic resonance imaging (MRI) demonstrated a multicystic tumor which had expanded within the left lobe of the liver and compressed the right anterior glisson sheath to the right. A mirror im-

Key words: Schwannoma, Liver, Neurofibromatosis

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age was seen in the cystic lesion, indicating that hemorrhage had occurred within it. Selective angiography (SAG) via the common hepatic artery showed that the left hepatic artery was distended but not encased by the tumor. Portography from the superior mesenteric artery demonstrated that the main trunk of the portal vein was shifted to the right by the large mass in the left lobe. Biochemical analysis, blood cell count and coagulation test were normal. Tumor markers in blood, including al-

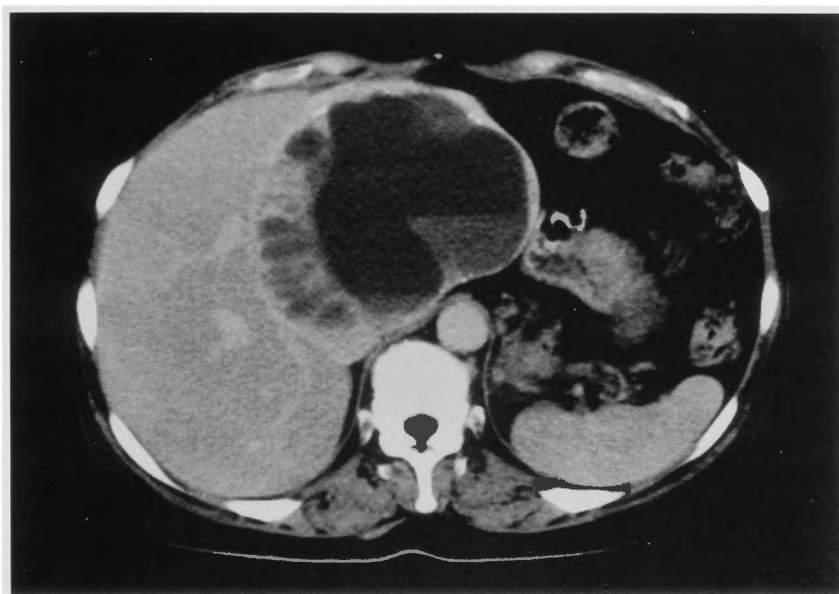


Fig. 1. Computed tomography of the liver. Large cystic tumor is located in the left lobe of the liver.

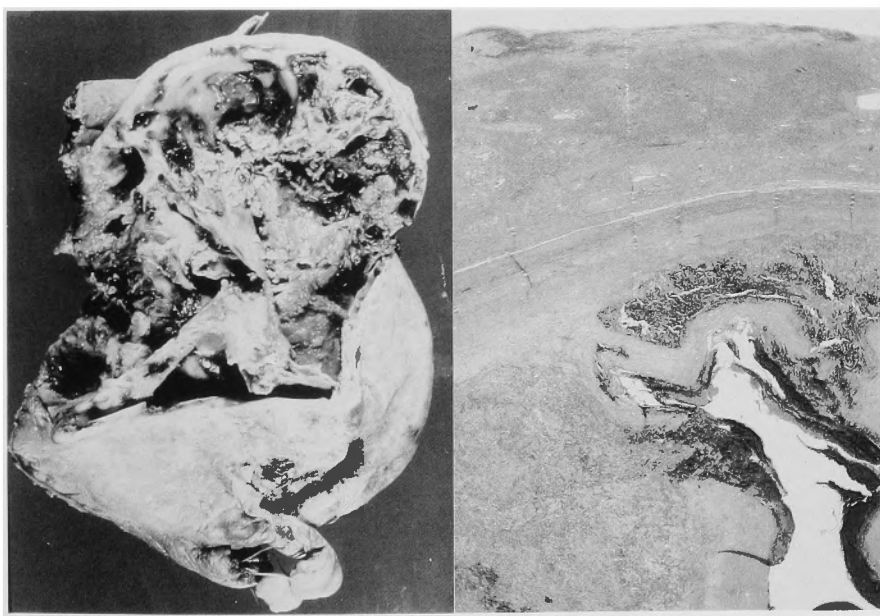


Fig. 2-A (left) and 2-B (right).

2-A. Gross appearance

2-B. Microscopic appearance. In low-power view, nonneoplastic liver parenchyma intervenes between peritoneal surface and fibrous capsule of the tumor (H & E, original magnification $\times 12.9$).

phafetoprotein, carcinoembryonic antigen, CA 19-9 and CA 125 were within normal range. Hepatitis-related antigens and antibodies were negative. Under preoperative diagnosis of cystadenocarcinoma of the liver, extended left lobectomy of the liver and cholecystectomy were performed. Regional lymph node clearing was not performed, since regional lymph nodes around the hepatoduodenal ligament along common hepatic artery and retroperitoneum were not enlarged in gross inspection. The post-operative course was uneventful and the patient was discharged on the 35th

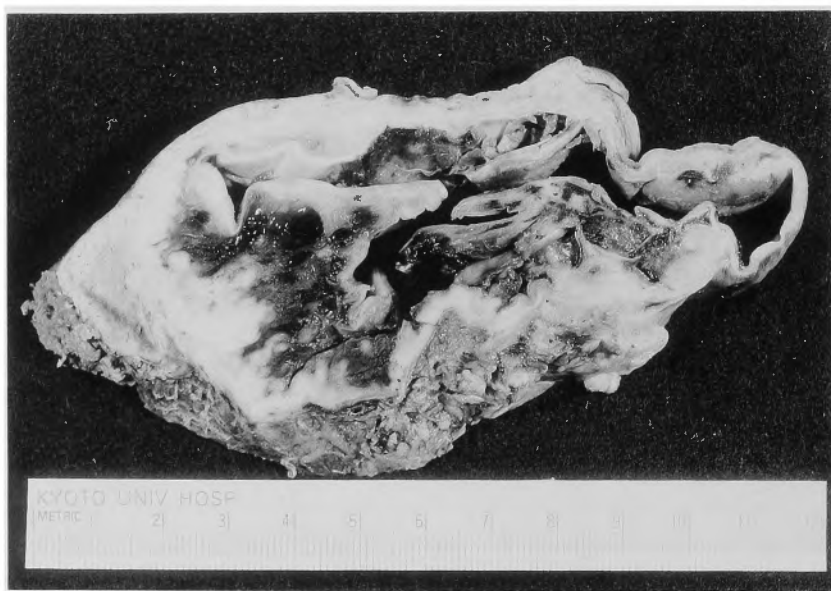


Fig. 3. Cutting surface of the tumor. Cystic change and hemorrhage are prominent

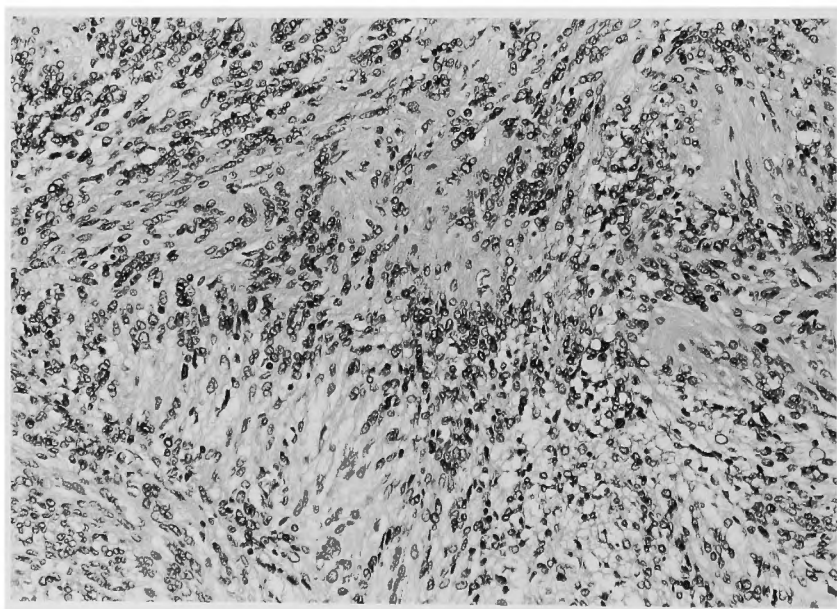


Fig. 4. Microscopic appearance. Proliferation of spindle-formed cells with nuclear palisading (H & E, original magnification $\times 257$)

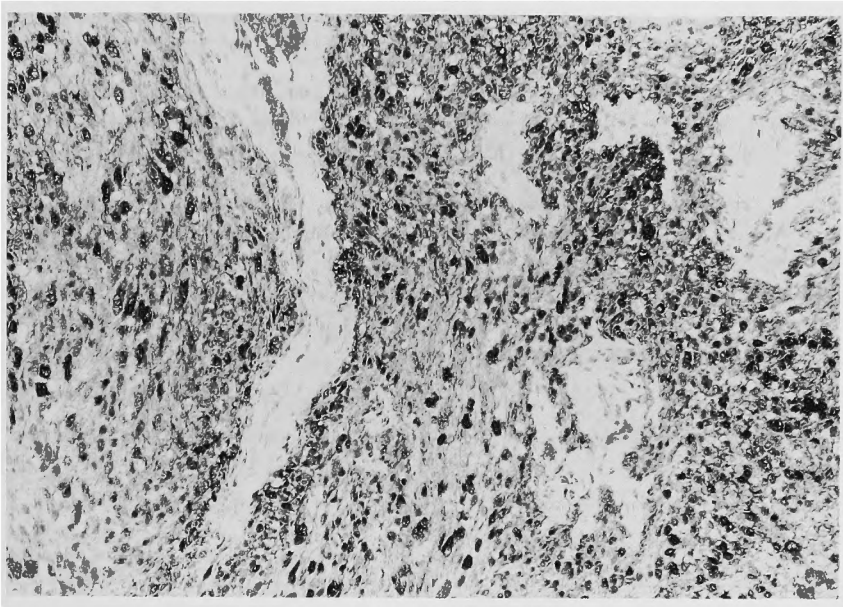


Fig. 5. Histochemical examination. S100 protein is intensely positive in the tumor cells. (Immunoperoxidase, original magnification $\times 257$)

postoperative day.

The surgical specimen consisted of an encapsulated mass of $16 \times 11 \times 13$ cm in size with adjacent small normal liver parenchyma. No distinct findings were seen in the gallbladder. On cutting the surface of the tumor, it contained a bloody fluid and pulverulent material with hemorrhage and showed features of cystic degeneration (Fig. 2A and 3). Microscopic examination showed a typical appearance of benign schwannoma with extensive cystic degeneration and hemorrhage (Fig. 4). Two different patterns could be recognized as Antoni A and B areas. The type A areas was quite cellular, composed of spindle cells arranged in palisading fashion. Mitoses were not recognized. In immunohistochemical stains, the S100 protein (DAKO, Glostrup, Denmark) was strongly positive both in the cytoplasm and nucleus of tumor cells (Fig. 5), while α smooth muscle actin (DAKO, Glostrup, Denmark) and desmin (DAKO, Glostrup, Denmark) were negative. The tumor was a bonafide intrahepatic lesion, and nonneoplastic liver parenchyma were interposed in most foci between the hepatic capsule and the tumor (Fig. 2B).

Comment

Of the various types of benign and malignant mesenchymal tumors affecting the liver, hemangioma is the most common benign one, while neurogenic tumors of the liver are very rare, irrespective of presence of neurofibromatosis⁴. Table 1 summarizes the hepatobiliary involvement in neurofibromatosis (von Recklinghausen's disease) in literature⁵⁻⁷. Two of these five cases were primary malignant schwannoma of the liver. Table 2 summarizes 6 cases of neurogenic tumors occurring in the liver of patients without neurofibromatosis in literature^{4, 8-11}. Of these 6, one was malignant, four were benign and the other was described as "semimalignant".

The innervation of the liver is by a hepatic plexus containing sympathetic and parasympathetic

Table 1. Hepatobiliary Involvement in von Recklinghausen's Disease

Source, y	Sex/Age	Location	Histological Diagnosis	Comment
Young SJ 1975 ⁽⁵⁾	M/23	Both lobes of the liver	Malignant schwannoma	Jaundice presented No metastase Autopsy performed
Mayer GW et al. 1974 ⁽⁶⁾	M/61	Ampulla of Vater	Neurofibroma	Obstructive jaundice presented Diagnosis made after resection of the tumor
Mayer GW et al. 1977 ⁽⁶⁾	F/37	Ampulla of Vater	Probably ganglioneuroma	Obstructive jaundice presented Exploratory laparotomy performed
Mayer GW et al. no date ⁽⁶⁾	F/53	Ampulla of Vater	Carcinoid	Diagnosis made by the intraoperative frozen section Tumor resection performed
Lederman SM et al. 1987 ⁽⁷⁾	M/21	Liver	Mixed type of malignant schwannoma and angiosarcoma	Autopsy performed Pulmonary metastases consisted of angiosarcomatous elements alone

Table 2. Neurogenic Tumors of the Liver Occurring in Patients without Neurofibromatosis

Source, y	Sex/Age	Histological Diagnosis	Comment
Shmurun RI and Chibisov VN 1977 ⁽⁸⁾	M/68	Malignant neurinoma	Autopsy performed Pulmonary metastases It is suspected that authors use the word "neurinoma" as schwannoma.
Pereira Filho RA et al. 1978 ⁽⁹⁾	F/56	Benign neurilemmal tumor	Biopsy performed in laparotomy Contains cystic nodule Whether it is neurilemoma or neurofibroma is not mentioned.
Bekker GM 1982 ⁽¹⁰⁾	M/70	Neurofibroma	Autopsy performed
Tuder RM and Moraes CF 1984 ⁽¹¹⁾	M/74	Semimalignant schwannoma	Subtotal hepatectomy performed, followed by hepatic insufficiency and death 21 days after surgery No metastase
Hytioglou P et al. 1993 ⁽⁴⁾	M/61	Benign schwannoma	Tumor resection performed Encapsulated tumor (13cm) with areas of hemorrhage, necrosis and cystification in the right lobe Half of the tumor was within the liver.
Yoshida M et al. 1993	F/56	Benign Schwannoma	Extended left lobectomy performed Encapsulated tumor (16cm) with areas of cystic degeneration and hemorrhage, containing pulverulent material All of the tumor was within the liver.

(vagal) fibers entering at the porta hepatis and largely accompanying the blood vessels and bile ducts; very few run among the liver cells and their terminals are uncertain. Both myelinated and non-myelinated fibers reach the liver from nerves in its various peritoneal folds³⁾. Therefore, it is possible that neoplasms originating from Schwann cells occur primarily in the liver.

The present tumor is considered to be a benign schwannoma of liver origin based on the following points of view. First, CT and MRI showed that the multicystic mass was located within the liver, and that normal liver parenchyma surrounded it. In the microscopic finding we could also verify that the liver parenchyma was continuously located between the tumor and the serosal surf-

ace. Second, the artery to the tumor originated from the left hepatic artery and during the operation we had to ligate many vessels passing through the liver parenchyma to the tumor.

Histological diagnosis of a benign schwannoma is usually a simple procedure in ordinary H & E section, and immunohistochemical staining for S100 protein is helpful for differential diagnosis of schwannoma from other types of spindle cell tumor including leiomyoma, leiomyosarcoma and fibrosarcoma¹³⁾.

Considering its size and form on the imaging, the tumor seemed to be malignant before operation. With the findings of multicystic lesion, large hemorrhage, and enhanced parenchyma around the cystic area, our preoperative roentgen diagnosis was cystadenocarcinoma of the liver. Although it is difficult to estimate the doubling time of the tumor, we suspected tumor growth accompanied with hemorrhage and cystic change. These findings are compatible with the first report of benign schwannoma of the liver irrespective of tumor size⁴⁾.

Many kinds of tumors in the liver show up as space occupying lesions (SOL) on films, such as CT and MRI. It is possible that a schwannoma can occur in the liver as one of these SOL, though it is very rare. Because the prognosis of benign schwannoma without neurofibromatosis is good, treatment of choice should be simple resection of the tumor, if preoperative diagnosis is confirmed. In the current case, however, we performed extended left lobectomy under preoperative diagnosis of cystadenocarcinoma without any postoperative complication since the hepatic function of our patient was normal and since the left lobe of the liver was atrophic due to tumor growth.

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和文抄録

肝臓原発の良性神経鞘腫

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肝臓に発生する神経原性腫瘍は神経線維腫症 (von Recklinghausen 病) の有無にかかわらず, 稀な疾患である。我々は, 56歳の神経線維腫症でない女性の肝臓

内に発生した良性神経鞘腫の症例を経験したので, 画像診断, 病理学的検査を含めて報告する。